Familial Mediterranean Fever Patient with Onset of Attacks at the Age of Seventies

Yetmişli Yaşlarda Atakları Başlayan Ailesel Akdeniz Ateşi Olgusu

Elderly Onset FMF

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Özet

Ailesel akdeniz ateşi (AAA) kendi kendini sınırlayan ateş, peritonit, plevrit ve/veya poliartrit atakları ile kendini gösteren ve otozomal resesif kalıtılan bir hastalıktır. Bir çocukluk çağı hastalığı olan AAA'de vakaların %80-90'ında tanı 20 yaşından önce konulmaktadır. Bu yazıda, 72 yaşında semptomları başlayan bir kadın AAA hastasını tanımladık.

Anahtar Kelimeler

Ailesel Akdeniz Ateşi; MEFV Gen Mutasyonu; Geç Başlangıçlı FMF; E148Q Mutasyonu

Abstract

Familial Mediterranean fever (FMF) is an autoinflammatory disease characterized by self-limited attacks of fever, peritonitis, pleuritis and/or polyarthritis, and autosomal recessive inheritance. 80-90 % of FMF cases are diagnosed before the age of 20 years. In this report, we described a female patient with FMF, symptoms starting at the age of 72 years.

Keywords

Familial Mediterranean Fever; MEFV Gene Mutation; Late Onset FMF; E148Q Mutation

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Introduction

Familial Mediterranean fever (FMF) is an autoinflammatory disease characterized by self-limited attacks of fever, peritonitis, pleuritis and/or polyarthritis, and autosomal recessive inheritance [1]. Mutations in MEFV, the gene encoding for marenestrin/pyrin is responsible for the disease [2]. In about 50 % of cases, attacks start in the first decade of life [3]. The mean age for starting of the symptoms is 4.5 years [4]. 80-90 % of FMF cases are diagnosed before the age of 20 years [3]. Late onset FMF (over 40 years of age) has been reported to have milder clinical outcome than early onset FMF and homozygous mutation of M694V has never been seen in this form [5]. In this report, we described a female patient with FMF, symptoms starting at the age of 72 years.

Case Report

A 73-years old female patient was admitted with recent onset of abdominal pain, fever, and arthritis of the knee. She was complaining of attacks of abdominal pain for one year and fever and arthritis were accompanying to abdominal pain for a few months. Her abdominal pain was lasting for about 3 days. Her son has been learned to have Behcet's disease and her nephew also has FMF. On physical examination, she had 38 °C fever, abdominal tenderness and arthritis of the right knee. She denied having recent tonsillitis or urinary tract infection. Laboratory analysis revealed that fibrinogen level was 627 mg/dl (200-400), erythrocyte sedimentation rate (ESR) was 88 mm/h and C-reactive protein (CRP) was 3.4 mg/L (0-4.9). Rheumatoid factor, antinuclear antibody serum anticyclic citrullinated peptide antibody levels were all negative. Brucella agglutination test was also negative. Proteinuria was absent in 24-hour urinary analysis. Plain radiography of the knee was not compatible for crystal arthropathy. Genetic analysis demonstrated heterozygous E148Q mutation. The patient was diagnosed as FMF and colchicine 1mg/day and meloxicam 15 mg/day were prescribed. With this treatment, symptoms of the patient were resolved. At follow-up, fibrinogen level was decreased to 438 mg/dl, ESR was 33 mm/h and CRP was 3.2. Abdominal pain, fever and arthritis were completely resolved.

Discussion

The age of FMF onset varies. The first attack occurs before the age of 10 years in about 60 % of FMF patients, before 20 years in 90 % and before 40 years in most of the remaining [5]. Onset of the disease after the age of 40 (late onset FMF) is rare [4].

In a study, 20 late onset FMF patients were evaluated. In this study, the frequency of late onset FMF was found as 0.5 %. Symptoms in late onset disease were mild [5]. None of the patients had M694V homozygosity which causes an earlier disease onset, a more severe course, more joint involvement, higher frequency of erysipelas-like erythema, higher doses of colchicine to control the disease [6] or chronic or prolonged manifestations of FMF such as amyloidosis, chronic arthritis, or protracted myalgia [7]. Our patient also has a mild disease without any amyloidosis and responded well to colchicine treatment.

In another study from Turkey, Sayarlioglu et al reported that among 401 FMF patients, 57 (14 %) were adult onset FMF (disease onset over 20 years of age) and 5 of them (2.25 %) had the first attack after 40 years of age [8]. In the study of Ureten et al, among 260 patients, 77 (30 %) were adult onset FMF but none of the patients were late onset FMF [9].

In our series, among 150 patients diagnosed as FMF, mean age for diagnosis was 24 years, 112 patient (74.6 %) had early onset disease, 38 patients (24.6 %) had adult onset disease. Only one patient (0.8 %) had the disease onset after the age of 40 (Erten S, Karakayalı A, unpublished data).

To the best of our knowledge, this is the first case of FMF with symptoms starting after the age of seventy-two. Adult-onset FMF may be a form of disease with distinct clinical, demographic, prognostic and molecular characteristics. Prospective clinical studies are needed to identify the characteristics of this phenotypic variant.

Competing interests

The authors declare that they have no competing interests.

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